

NEUROTOLOGIC EVALUATION AND MANAGEMENT OF ACOUSTIC NEUROMA*

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DIAGNOSIS of an acoustic neuroma depends upon a high index of suspicion and subsequent thorough laboratory evaluation. Cushing,¹ in his 1917 monograph, *Tumors of the Nervus Acousticus*, first emphasized tinnitus and hearing loss as early symptoms of an eighth cranial nerve tumor. Still, due to a lack of confirmatory tests, patients came to operation only with signs and symptoms of far advanced disease: papilledema, blindness, cerebellar ataxia, and multiple cranial nerve neuropathies. Cushing advocated a bilateral suboccipital craniotomy because of the difficulty in localizing the side of the lesion. He considered partial enucleation as essential to keep operative mortality in the range of 20%.

The advent of audiology, vestibular tests, and improved radiologic techniques opened a new diagnostic and therapeutic era. William House² in the 1960s popularized a full neurotologic evaluation for all patients with tinnitus, hearing loss, or dizziness. Emphasis shifted toward the earlier diagnosis of small tumors. The translabyrinthine surgical approach to the cerebellopontine angle, using microsurgical technique, allowed for complete tumor removal with preservation of normal facial nerve function. Operative mortality fell to less than 5%. Most patients were able to return to their preoperative work and life style. Further recent advances, such as the middle cranial fossa surgical approach to the temporal bone, have allowed preservation of hearing in patients with small tumors.

DIAGNOSIS

The key to the diagnosis of an acoustic neuroma is a high index of suspicion. Any patient whose signs or symptoms suggest a cerebellopon-

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tine angle lesion should undergo a full neurotologic evaluation. The following is a brief overview:

History. The most common presenting symptom of an acoustic neuroma is progressive unilateral hearing loss with or without tinnitus. Some patients, due to a slow deterioration in hearing, are unaware of their hearing loss, and these patients may complain only of tinnitus or, often, of ear fullness. Ten percent of patients with a confirmed acoustic neuroma have first complained of a sudden loss of hearing.

Although an acoustic tumor usually arises from the vestibular division of the eighth cranial nerve, vertigo is unusual. Most patients describe only a vague sense of imbalance or lightheadedness. Headache, loss of coordination, facial numbness and loss of vision are signs of large tumors and advanced disease. Facial nerve paralysis is not common and suggests a facial nerve neuroma.

It bears emphasis that there is no typical history.³ Any of the aforementioned symptoms, alone or in combination, merit a full evaluation. Even a patient with the classical history of Meniere's disease must be considered suspect.

Physical examination. A routine otolaryngic examination of the head and neck should be performed on every patient. Cranial nerve function is of particular importance in evaluating the extent of disease. Corneal hypesthesia (first division of the trigeminal nerve) is often the first sign of tumor extension into the cerebellopontine angle. Cerebellar testing should evaluate rapid alternating movements (diadochokinesia) and fine coordination. The Romberg test may be refined as the sharpened tandem Romberg test (one foot in front of the other, arms crossed over chest, eyes closed).

Audiologic evaluation. Audiologic evaluation is mandatory for any patient with a hearing or balance complaint. Pure tone and speech discrimination are a minimum. Site of lesion testing, particularly stapedial reflex and reflex decay, help to identify eighth nerve lesions.

Vestibular evaluation. Because the acoustic neuroma usually arises from the balance portion of the eighth cranial nerve, it is important to evaluate vestibular function. Caloric tests should be performed with electronystagmography, which takes advantage of the corneoretinal dipole, and thus allows for precise recording of eye movement behind closed eyes. Utilizing electronystagmography, a reduced caloric response has been demonstrated on the side of the tumor in as high as 86% of patients.

Radiologic evaluation. Radiologic studies are fundamental to the



Fig. 1. Computerized axial tomogram with contrast enhancement demonstrating a large right cerebellopontine angle mass (arrows), surgically confirmed as an acoustic neuroma.

neurotologic evaluation. Routine petrous pyramid roentgenograms have been replaced by complex motion tomography, which allows detailed evaluation of the size and configuration of the internal auditory canals. Any enlargement of the internal auditory canal on the side of suspicion merits further study.

Computerized axial tomography is of great value in the diagnosis of

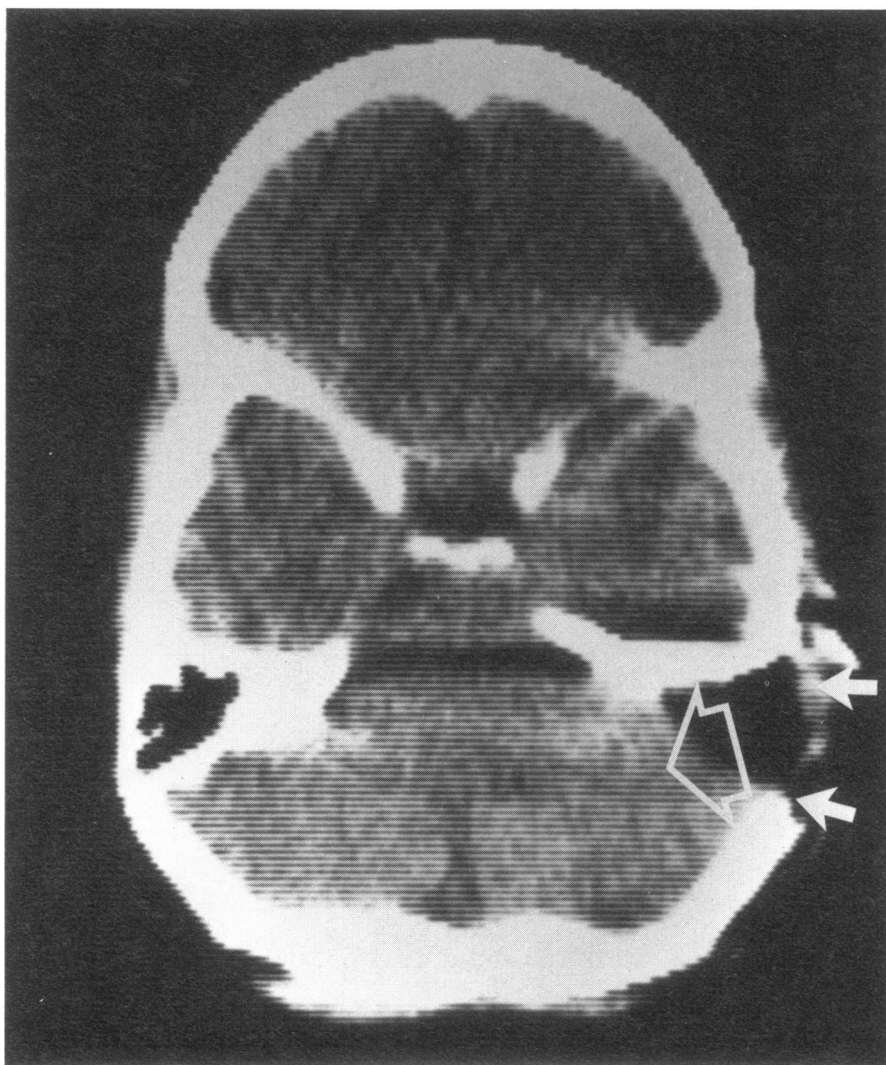


Fig. 2. Postoperative computerized axial tomogram demonstrating access to the cerebellopontine angle through the translabyrinthine approach.

cerebellopontine angle tumors. The acoustic neuroma is isodense, but can be enhanced by intravenous contrast media (Figure 1). An important limitation of computerized tomography, however, is its inability to detect a lesion less than 1.5-2.0 cm. in diameter in the cerebellopontine angle. A small acoustic neuroma, which can be removed with less morbidity and possible preservation of hearing, will not be detected by computerized tomography.

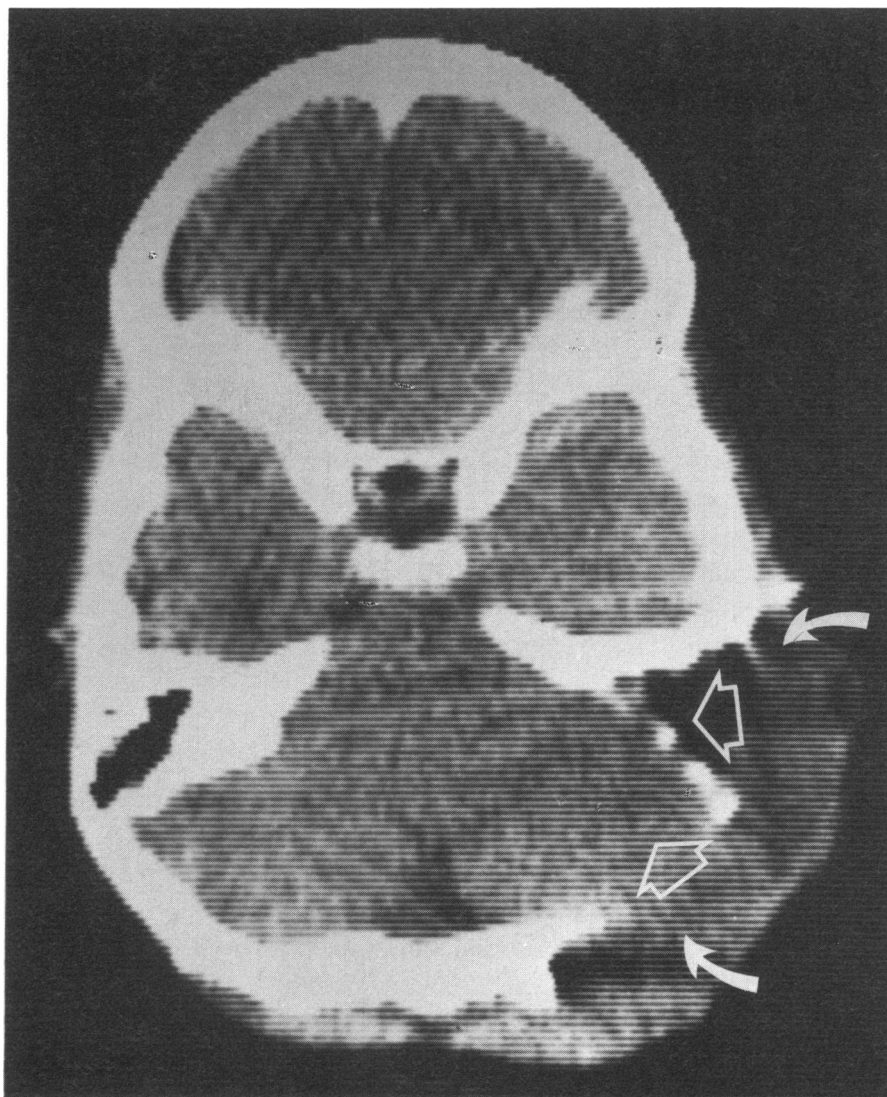


Fig. 3. Postoperative computerized axial tomogram (one week postoperative—the dressing is seen on the scan overlying wound) demonstrating access to cerebellopontine angle through the translabyrinthine approach with retrosigmoid extension.

Posterior fossa myelography is the most accurate radiologic study in the diagnosis of an acoustic neuroma. It may be performed with as little as 0.5 ml. of injected contrast material, and carries almost no morbidity. With rare exceptions, pneumoencephalography and arteriography are not indicated in the evaluation of the acoustic neuroma.

SURGICAL MANAGEMENT

The neurotologic approach to an acoustic neuroma is designed to allow total tumor removal with preservation of normal facial nerve function. All surgery is performed using an operating microscope.

Middle cranial fossa approach. The middle cranial fossa surgical approach is indicated for those patients whose tumors are limited to the internal auditory canal and who have useful hearing. This approach allows total tumor removal and preservation of hearing in 50% to 60% of patients.

Translabyrinthine approach. The translabyrinthine approach is indicated for most acoustic tumors (Figure 2). This approach allows complete tumor removal and offers the best chance of facial nerve preservation. Cerebellar retraction or resection is not necessary.

Combined approach (Figure 3). Large acoustic tumors (greater than 4 cm.) are exposed initially through a translabyrinthine approach. Most are completely removed through this approach. When anatomical limitations or vital signs changes preclude complete removal, a retrosigmoid dural flap is elevated at the same sitting. This allows for cerebellar retraction and additional exposure.

CONCLUSION

The importance of a thorough history and complete physical examination cannot be overemphasized. The challenge is to diagnose the acoustic neuroma when it is small, when it can be removed with little morbidity and no mortality, and when hearing may be preserved. Cushing's comment on the issue of a 25-year-old salesman with blindness, cerebellar ataxia, and multiple cranial nerve neuropathies is exemplary:

The significance of the initial auditory disturbances was not appreciated when this patient was seen, and it is not improbable that they were of long standing and accompanied by vertiginous attacks. These matters were not thoroughly inquired into, and the fact of his unilateral deafness was only casually mentioned among the notes of the physical examination.

Any hearing loss, regardless of its degree, merits a neurotologic evaluation. The key to diagnosis remains a high index of suspicion.

REFERENCES

1. Cushing, H. *Tumors of the Nervus Acousticus*. Philadelphia, Saunders, 1917.
2. House, W. editor: Monograph II. Acoustic neuroma. *Arch. Otolaryngol.* 88:715, 1968.
3. Hoffman, R. and Brookler, K. H.: Under-rated neurotologic symptoms. *Laryngoscope* 88:1127-37, 1978.